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Solid pseudopapillary tumour of pancreas: A rare tumour with good prognosis: A case report

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Abstract
Solid pseudopapillary neoplasm (SPN) of pancreas is a rare exocrine tumour of pancreas. It has low malignant potential with excellent prognosis and usually effects females in adolescent age group. We report a case of 40 year old female who presented with pain abdomen. Radiological investigations revealed a mass lesion in the pancreas. The diagnosis was confirmed on histopathological examination. Due to its low incidence, the clinical and pathological features of this tumour have not been extensively studied. It is important to be aware of its specific microscopic features, immunohistochemistry and differential diagnosis as timely surgical resection provides long- term survival.

Keywords: Solid pseudo papillary neoplasm, pancreas, exocrine tumour

Introduction
Solid pseudopapillary neoplasm (SPN) is a rare pancreatic tumor making less than 2% of all exocrine pancreatic tumors. Mostly patients are female in adolescent age group and in second or third decade of life. The tumors is mostly found coincidently on routine physical examination or imaging study and during abdominal trauma workup. It is a tumor with low malignant potential but surgical resection remains the main treatment.

Case report
A 40 year old female presented with pain abdomen since one month. There was no history of trauma, fever, vomiting, urinary or bowel symptoms or jaundice. No past history of chronic illness or significant family history. General physical examination revealed a healthy female. Review of systems was otherwise unremarkable. Further investigations were ordered and ultrasonography revealed bulky pancreas in body region showing an irregular cystic lesion measuring 24x16mm. Head and tail were normal in echotexture. CECT (Contrast Enhanced Computed Tomography) abdomen showed heterogeneously enhancing mass lesion in the neck of pancreas with dorsal agenesis. MRCP (Magnetic Resonance Cholangiopancreatography) was done and revealed an ill-defined mass in body of pancreas giving an impression of Solid pseudopapillary neoplasm with dorsal agenesis. USG (Ultrasonography) guided FNAC (Fine Needle Aspiration Cytology) revealed hemorrhagic smear. Tumor markers like carcinoembryonic antigen (CEA), carbohydrate antigen (CA19-9), and alpha-fetoprotein (AFP) were all within normal range. Spleen preserving pancreatic mass resection with cholecystectomy was done under general anesthesia and we received three specimens of common hepatic lymph node, gall bladder and pancreatic mass.
Grossly, pancreatic mass was grey white to grey brown measuring 4x3.5x2 cm. Cut section of pancreas showed 2 grey white areas, one measuring 2x1.3 cm, another measuring 0.6 cm in diameter and 2 cystic areas (Figure 1). Common hepatic lymph node and gall bladder specimens were grossly unremarkable.

Fig 1: Gross specimen of pancreatic mass showing solid grey white to grey brown areas with cystic degeneration

Microscopic examination of sections from pancreatic mass revealed well circumscribed tumor. Solid areas revealed tumor cells arranged in organoid pattern separated by thin fibrovascular septa, acini and sheets (Figure 2).

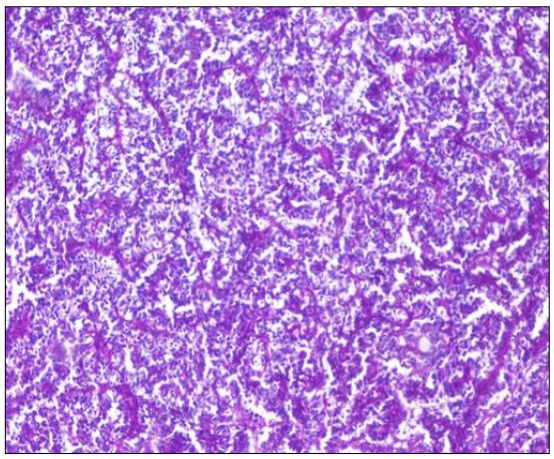

Fig 2: Microphotograph of solid areas show tumour cells arranged in organoid pattern separated by thin septa (H&E, 10x)

Individual tumor cells have round, oval, elongated nuclei, finely dispersed chromatin, inconspicuous nucleoli and moderate amount of eosinophilic cytoplasm (Figure 3). Some nuclei were having grooves/indentation. Prominent intracytoplasmic and extracellular hyaline globules (PAS +, diastase resistant) were also present (Figure 4A & 4B).

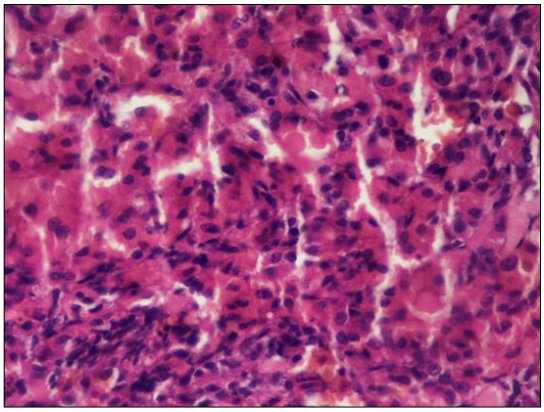

Fig 3: Microphotograph showing individual tumour cells with round to oval nuclei, finely dispersed chromatin, inconspicuous nucleoli and eosinophilic cytoplasm. Focal oncocytic change also seen (H&E, 40x)

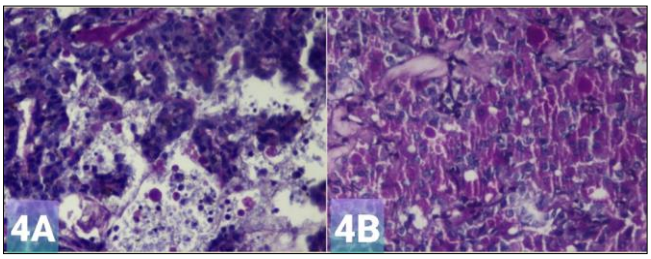

Fig 4A: Microphotograph showing intracytoplasmic and extracellular hyaline globules. Figure 4B: Microphotograph showing PAS positive hyaline globules

Tumor also revealed rich delicate vascular network and focal oncocytic change in tumor cells. Stroma showed foci of hyalinization and dystropic calcification. No mitotic figures were seen. Cystic areas revealed papillae (Figure 5) hemorrhage and marked fibrin deposition. Normal pancreatic acini and ducts were pushed to the periphery. Perineural invasion was also present.

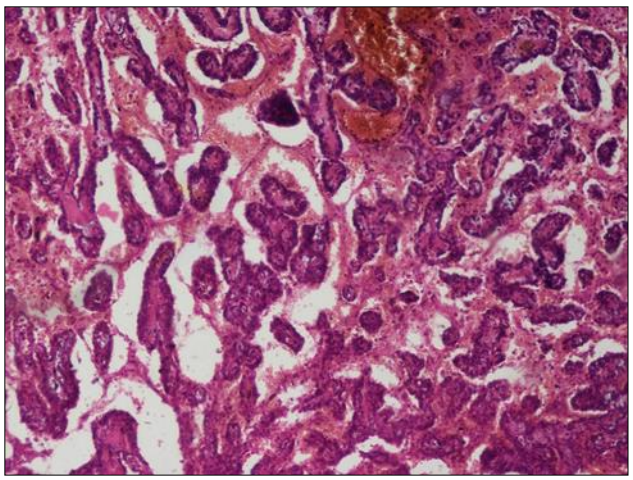

Fig 5: Microphotograph of cystic areas show papillae formation

Final diagnosis of solid pseudopapillary tumor (SPT) of pancreas was made.

Postoperative course was uneventful and no additional therapy was administered.

Discussion

Solid pseudopapillary tumor (SPT) of the pancreas is one of the rare exocrine pancreatic tumor, which accounts for 2 to 3% of all tumors of the pancreas and 0.9 to 2.7% of exocrine pancreatic neoplasms [1, 2]. It has low malignant potential and excellent prognosis [3]. It was first described by Frantz in 1959 [4]. Different names were reported for this tumour until it was defined by the World Health Organization (WHO) in 1996 as a “solid pseudopapillary tumor” of the pancreas (5). The pathogenesis of this tumour is unclear. Many investigators favor the theory that SPTs originate from multipotent primordial cells, whereas others suggest the possibility of extrapancreatic origin [6].

This tumor has a predilection for young Asian and African-American women. The male to female ratio is 1:10 and the mean age at presentation is 22 years. However, it can also effect pediatric population. Clinically, it is often asymptomatic or present with gradually enlarging abdominal mass and pain which is the most common presenting clinical symptom or sign. Other non specific symptoms include nausea, vomiting, fever, weight loss and jaundice. Many of them are caused due to tumour

compression of normal pancreas [7]. However, due to its low incidence, its clinical and pathologic features have not been extensively studied. Although most SPTs exhibit benign behavior, malignancy can occur in about 15% of cases, manifesting as metastases or invasion of adjacent structures [6]. Most common metastatic sites were omentum and liver [8]. The most common site is pancreatic body and tail followed by head and neck. However, even if the tumour was located at the head of pancreas, obstructive jaundice was very rare due to the tumour exophytic growth way, which was consistent with few studies [9].

The gross appearance of SPT varies from solid to cystic components with cellular degenerative changes. The tumour is well capsulated and demarcated from the pancreas [10]. They are characteristically positive for alpha 1-antitrypsin, beta catenin, CD56, CD10, and vimentin. In addition, other positive immunohistochemical markers include alpha-1 antichymotrypsin (AAT), cyclin D1 and synaptophysin [3]. Although there is distinct female preponderance for SPN, estrogen receptor positivity is very uncommon but on the other hand, progesterone receptor positivity is seen in almost all cases of SPN, irrespective of sex. Keratin positivity is seen in 30% to 70% of cases [11].

The majority of tumors are diagnosed through CT (Computed tomography) and MRI (Magnetic Resonance Imaging) of the abdomen. Plain radiography (X- ray) is not of much help and reveals possible calcification in the tumour. CT scan of the tumour show solid and cystic areas with regions of hemorrhage and focal calcification. Lee *et al.* Reported that pancreatic duct dilation and vessel invasion, either with or without metastasis points towards aggressive behavior of this neoplasm [12]. MRI also defines the hypervascular, well-encapsulated, round tumors with mixed cystic and solid components with high and low signal intensity on T1 and T2 weighted images. Calcification may be present in few cases, whereas dilation of pancreatic duct is rare. Echo-endosonography may provide FNA biopsy with the possibility of pre-operative pathologic diagnosis. Incomplete capsule which means a capsule that did not surround the entire periphery of the tumour radiologically corresponds to infiltrative growth pattern microscopically in most of the cases. This infiltrative growth pattern might cause disruption of capsule and indicate malignant behavior. The tumor has a low-grade malignant potential and tends to have a favorable prognosis, even in the presence of metastatic disease [13].

Grossly, these neoplasms widely vary in size. They measure between 0.5 cm to 34.5 cm with a mean of 9.3 cm. The tumour is usually a well circumscribed round mass separated from the pancreatic parenchyma by a pseudocapsule. Cut section of the tumour show solid and cystic areas along with hemorrhage and necrosis. Incomplete capsule during gross examination is highly suggestive of a malignant SPT. The larger the tumour, the greater is the cystic component [14].

Histopathological features represent combination of solid and cystic components. Solid component shows pseudopapillae with tumour cells revealing uniform centrally placed round to oval small nuclei with grooves and moderate amount of amphophilic cytoplasm with focal aggregation of intracytoplasmic and extracytoplasmic hyaline globules. These globules are typically periodic – acid – schiff positive and diastase resistant and is highly characteristic for diagnosis of SPN. The nuclei do not have salt and pepper features, which is diagnostic of neuroendocrine tumours. Foamy macrophage and foreign body giant cells are commonly observed adjacent to cystic spaces. Mitotic activity is minimal or absent [15].

Microscopic features of high grade malignancy in aggressive SPN includes diffuse growth pattern with minimal fibrovascular stroma, extensive tumour necrosis in diffuse, geographic or punctate pattern, increased nuclear to cytoplasmic ratio with hyperchromasia and a high mitotic index (upto 70 mitosis per 50 high power fields) [16]. SPN are considered to be low grade malignant neoplasms, so finding positive lymph nodes is very uncommon. However few cases of aggressive fast growing SPN have reported liver and lymph node metastasis.

Overall 5-year survival is as high as 97% in patients undergoing surgical resection [17]. Neither vascular, lymphatic or perineural invasion has been a factor for predicting tumor recurrence or overall survival of patients. Surgery is the treatment of choice, even in the case of distant hepatic metastasis or local recurrence. Type of operation depended on tumour's size, location and intraoperative frozen section examination. Due to the low malignant nature of the tumour, organ- preserving operation should be performed whenever feasible [18].

Differential diagnosis of SPN includes pancreatic neuroendocrine neoplasms, acinar cell carcinoma, pancreatoblastoma and serous cystadenoma. Pancreatic neuroendocrine tumours are very similar to SPN and is often difficult to diagnose alone by histology or cytology. Histological features like presence of pseudopapillae, foamy histiocytes, hyaline globules and nuclear grooving favor SPN. On the other hand, presence of salt and pepper chromatin favors neuroendocrine tumour. Immunohistochemistry is recommended to differentiate between these two entities in most cases. Cohesive clusters of acinic cells and with abundant acinar formation having granular cytoplasm, irregularly sized nuclei maintaining cytoplasmic polarity is the key for diagnosis of acinar cell carcinoma [19]. Pancreatoblastoma is a rare pancreatic tumour that mainly occurs in children and histologically shows epithelial cells surrounded by a fibrous stroma. The epithelial component consists of cells arranged in acini, sheets and squamoid corpuscles. Squamoid corpuscles and nuclear clearing are diagnostic features of pancreatoblastoma [20]. Elevated alpha fetoprotein also indicates towards diagnosis of pancreatoblastoma. Serous cystadenoma is a circumscribed nodule consisting of cystic spaces lined by cuboidal cells with clear cytoplasm containing glycogen. Myoepithelial layer is present and cell islets are present between lobules, forming a radiating pattern with central scar [21].

Conclusion

It is essential for a pathologist to know that SPN is rare tumour of pancreas with uncertain origin, particularly in females. It is a tumour of low malignant potential and good prognosis. It is important to be familiar with its unique microscopic features, immunohistochemical panel and differential diagnosis, in particular neuroendocrine tumour. Aggressive surgical resection ensures cure in majority of patients [22].

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